AMSER Case of the Month September 2025

2-month-old male with vomiting and failure to thrive.

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Patient Presentation

2-month-old male, former premature twin gestation (36 weeks, SGA) who presented with failure to thrive, poor weight gain, poor feeding, and non-bloody, non-bilious emesis. Pyloric stenosis was suspected.

What Imaging Should We Order?

Applicable ACR Appropriateness Criteria

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Variants

1. Infant older than 2 weeks and up to 3 months old. New onset nonbilious vomiting (suspected hypertrophic pyloric stenosis). Initial imaging.

Documents

Documents

- Narrative
- Evidence Table
- Lit Search
- Appendix

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Vomiting, nonbilious, new onset, hypertrophic pyloric stenosis suspected, initial imaging	3074079	US abdomen (UGI tract)	0 mSv O	0 mSv [ped] O	Usually appropriate
		Fluoroscopy upper GI series	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	May be appropriate
		Radiography abdomen	0.1-1mSv ☢☢	0.03-0.3 mSv [ped] ☢☢	Usually not appropriate
		Fluoroscopy contrast enema	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate
		Nuclear medicine gastroesophageal reflux scan		0.3-3 mSv [ped] ☢☢☢	Usually not appropriate

Ultrasound was ordered by the pediatrician

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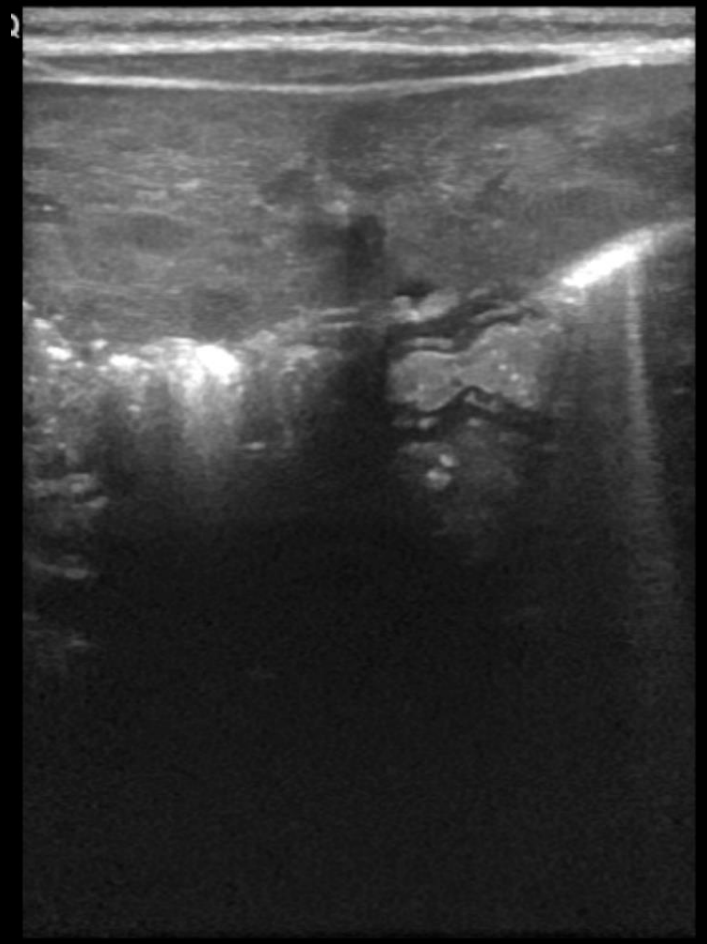
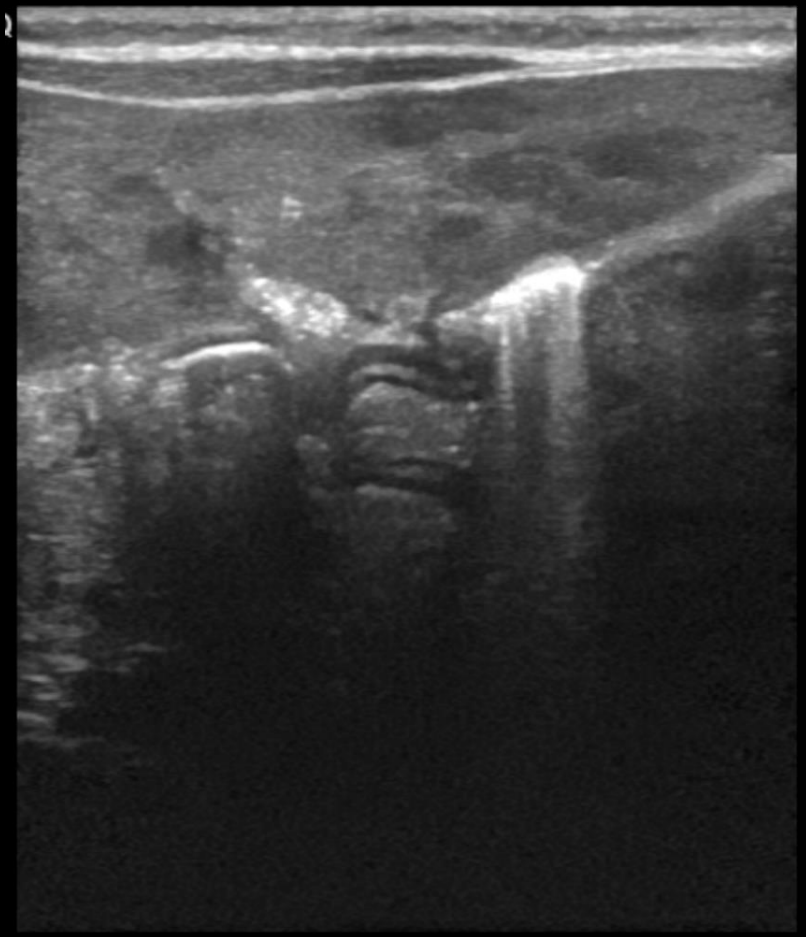
Findings (unlabeled)





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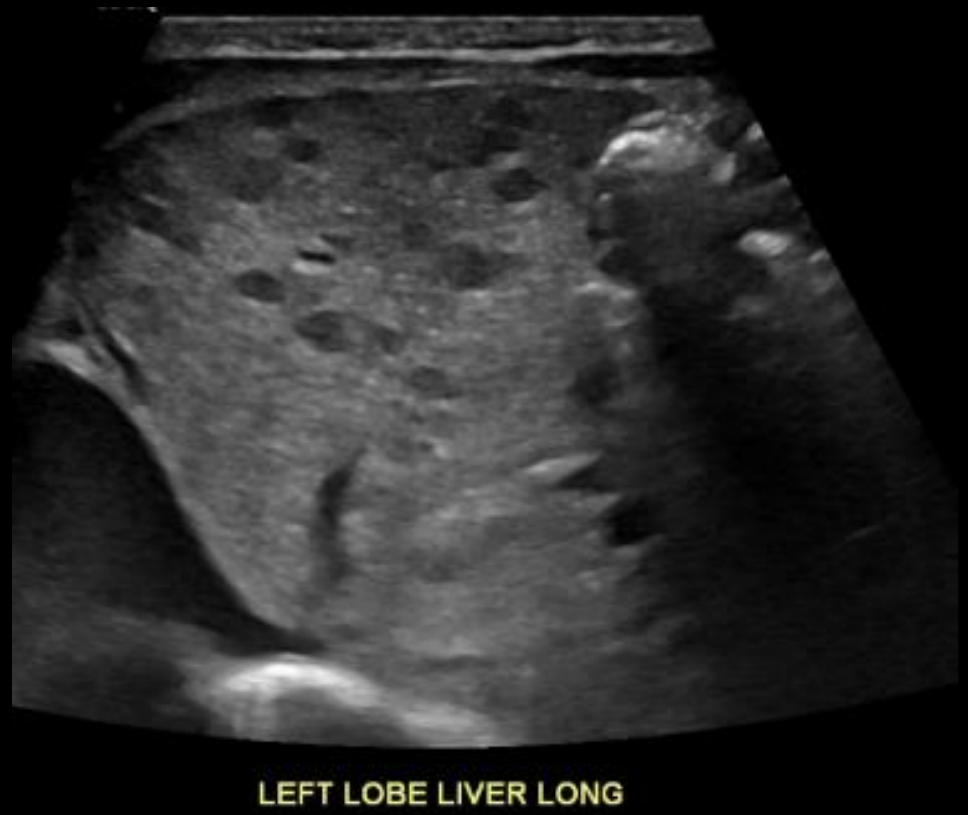
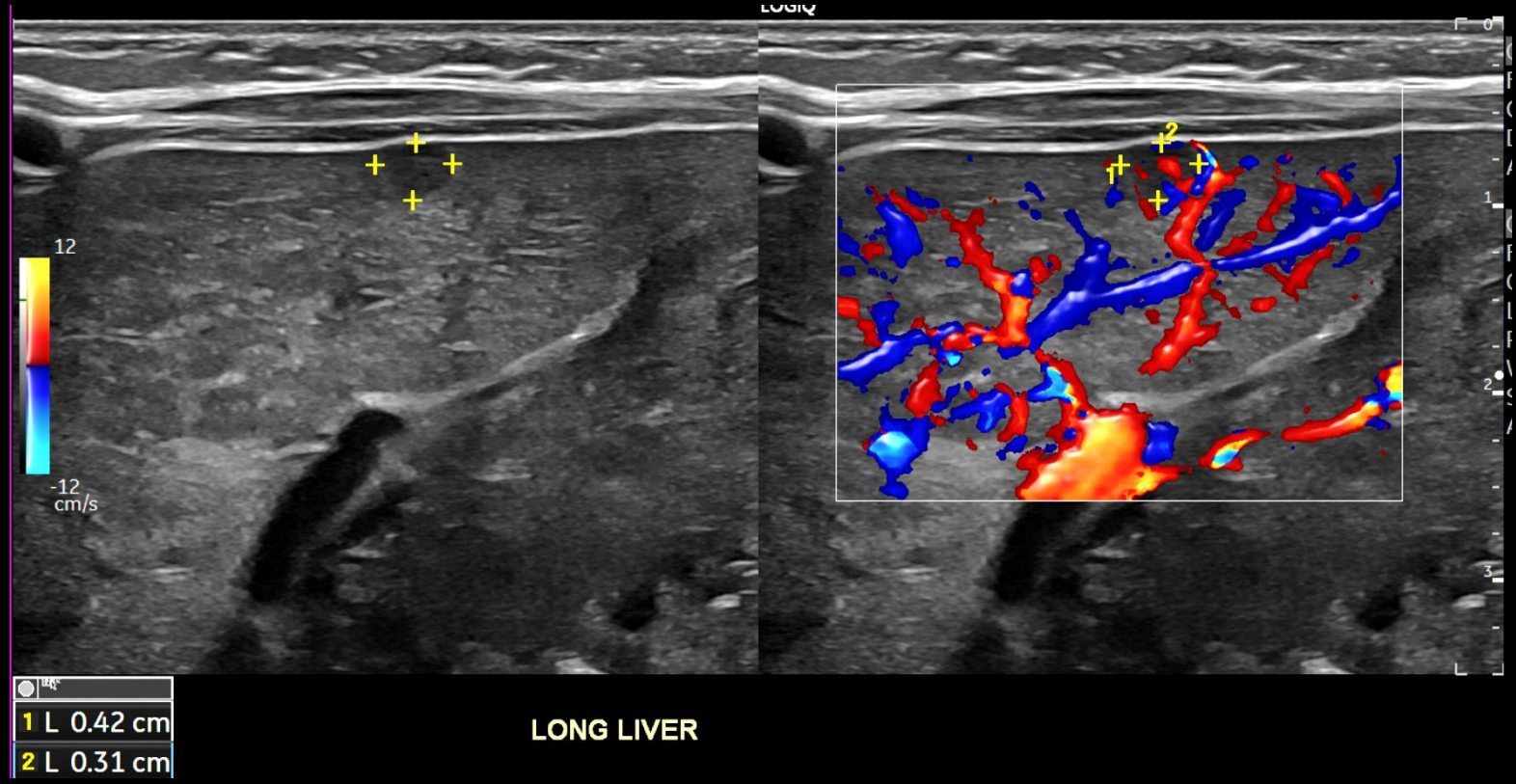
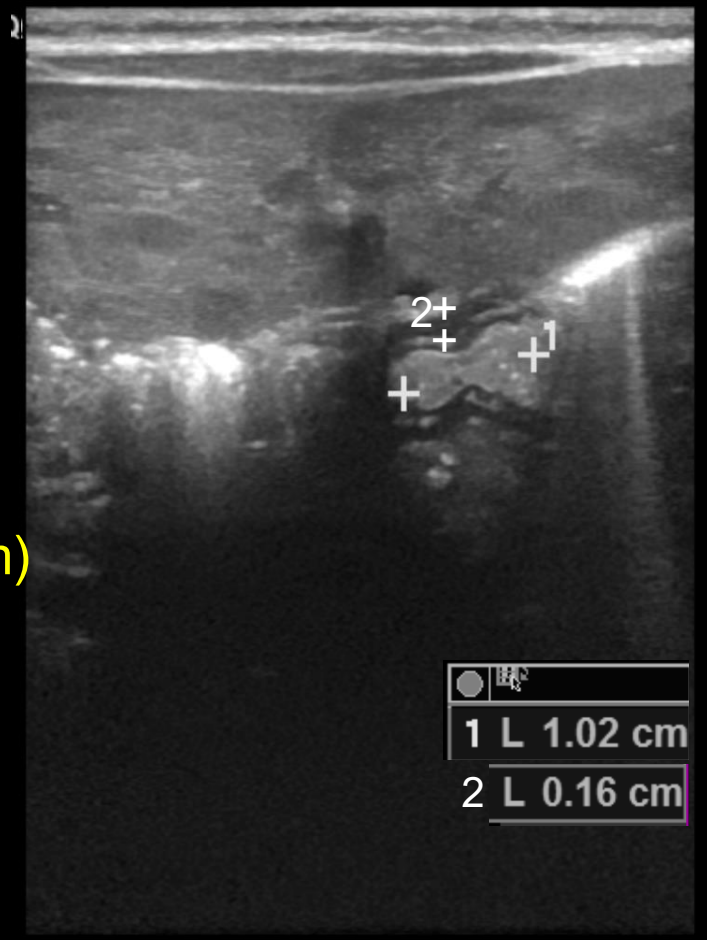
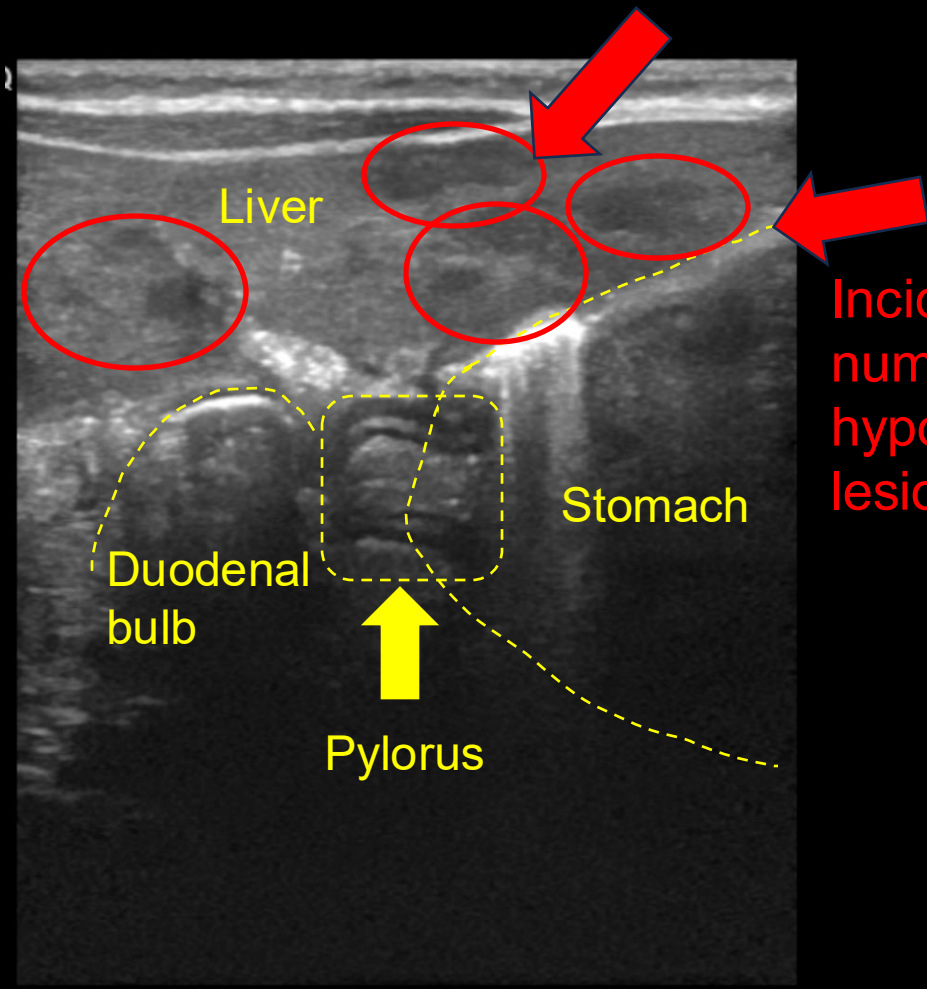




Findings (labeled)

Normal pylorus channel length (<1.6 cm) and normal wall thickness (< 3 mm)





Incidental finding of numerous small hypo-echogenic liver lesions

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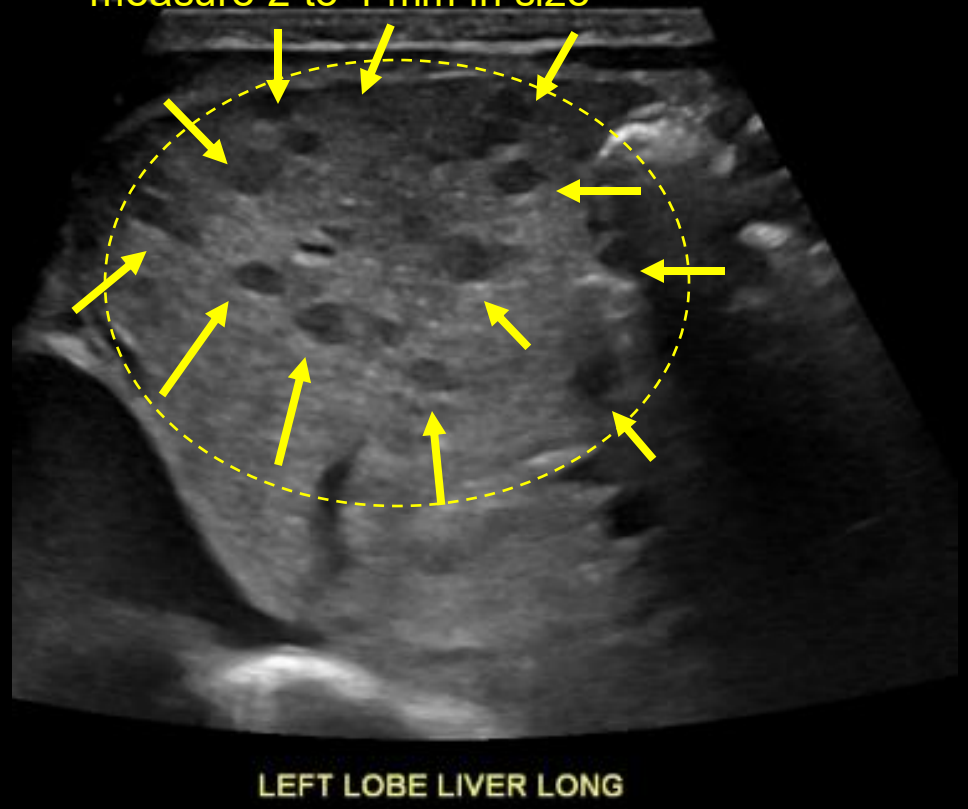
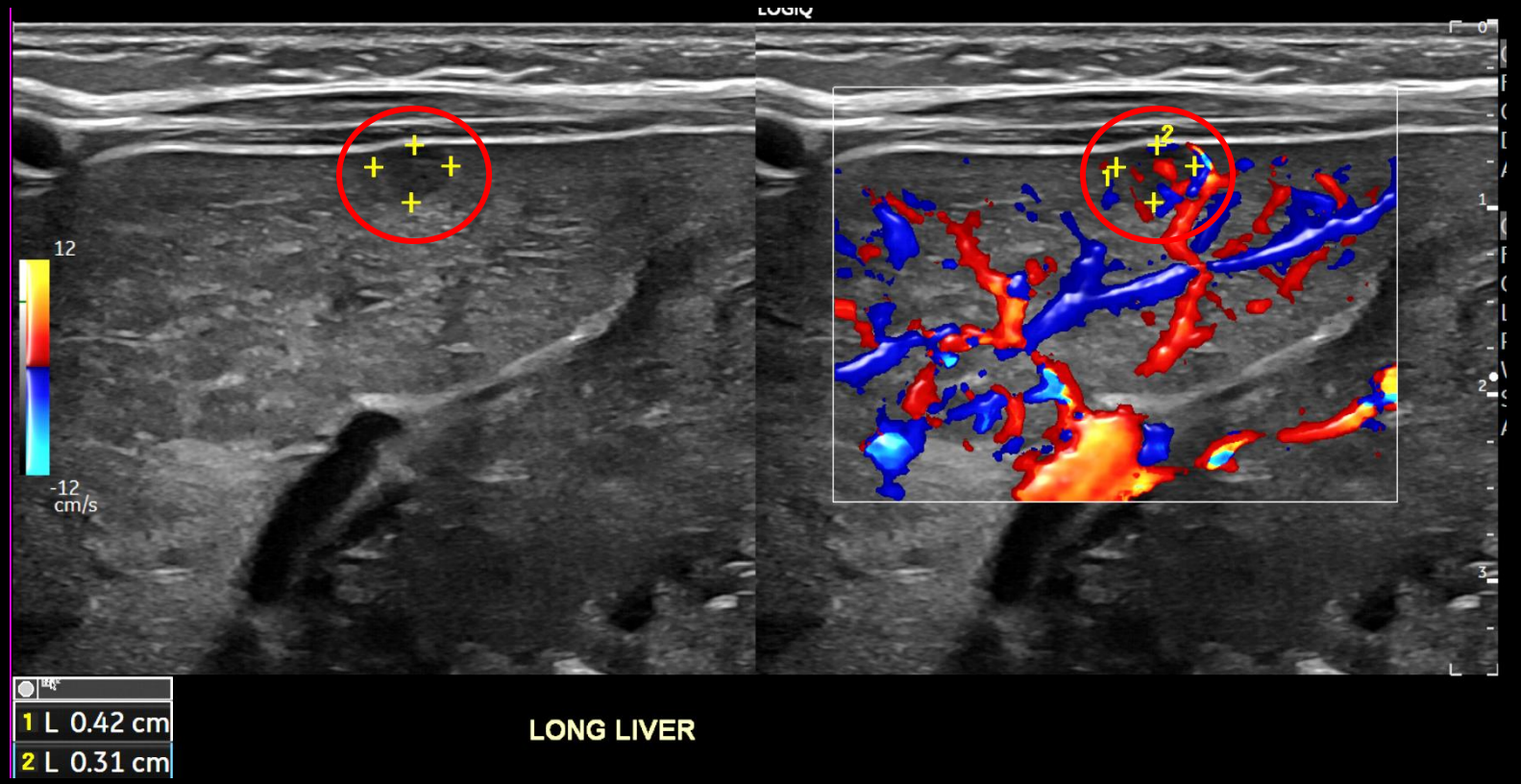
Findings (labeled)

Numerous hypoechoic lesions randomly distributed diffusely throughout the liver. The lesions are well-circumscribed and measure 2 to 4 mm in size



Focused side-by-side grayscale and color Doppler imaging demonstrates hypervascular nature of the small hypoechoic lesions



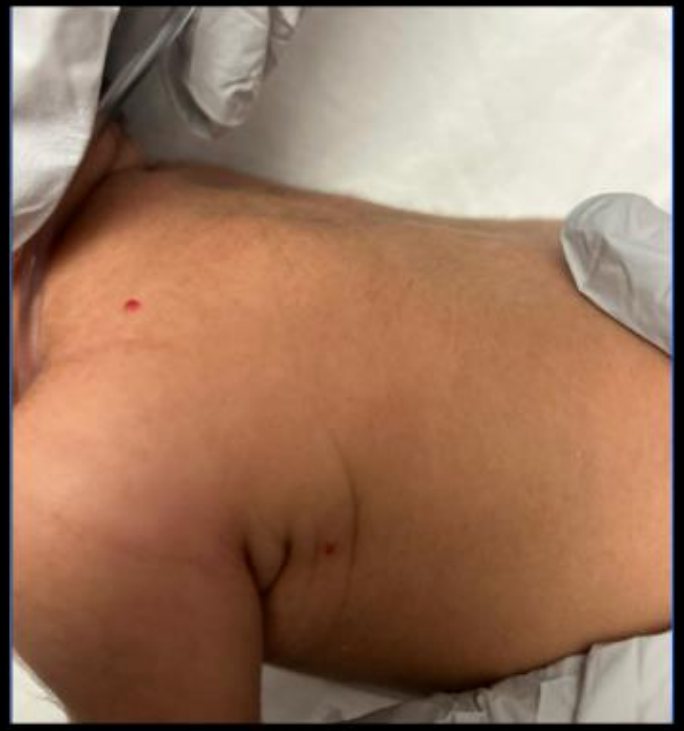
Additional diagnostic work-up was pursued

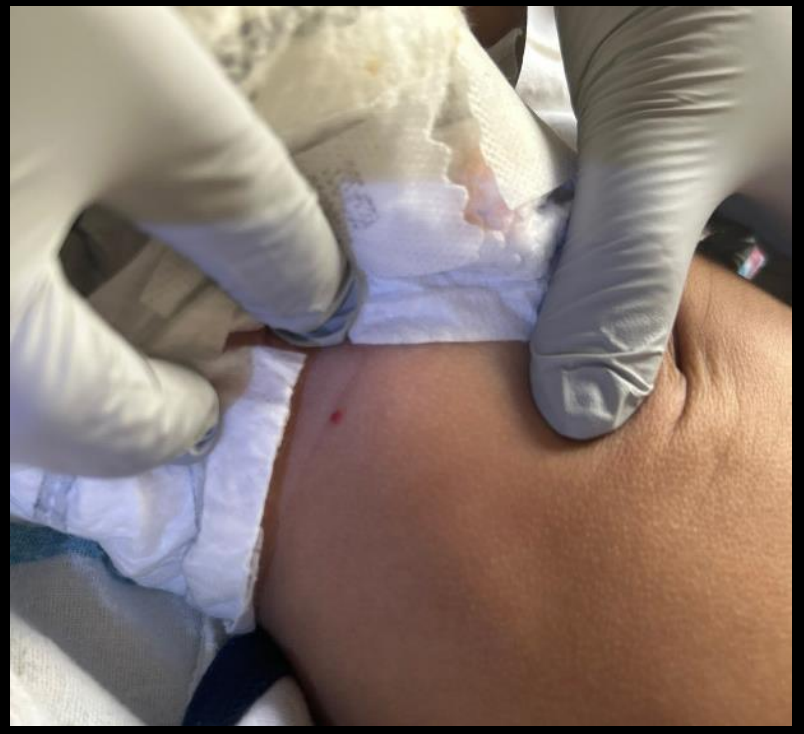
Laboratory data
MRI of the abdomen and pelvis
Echocardiogram
Bone survey
Genetics profiling

Pertinent Labs

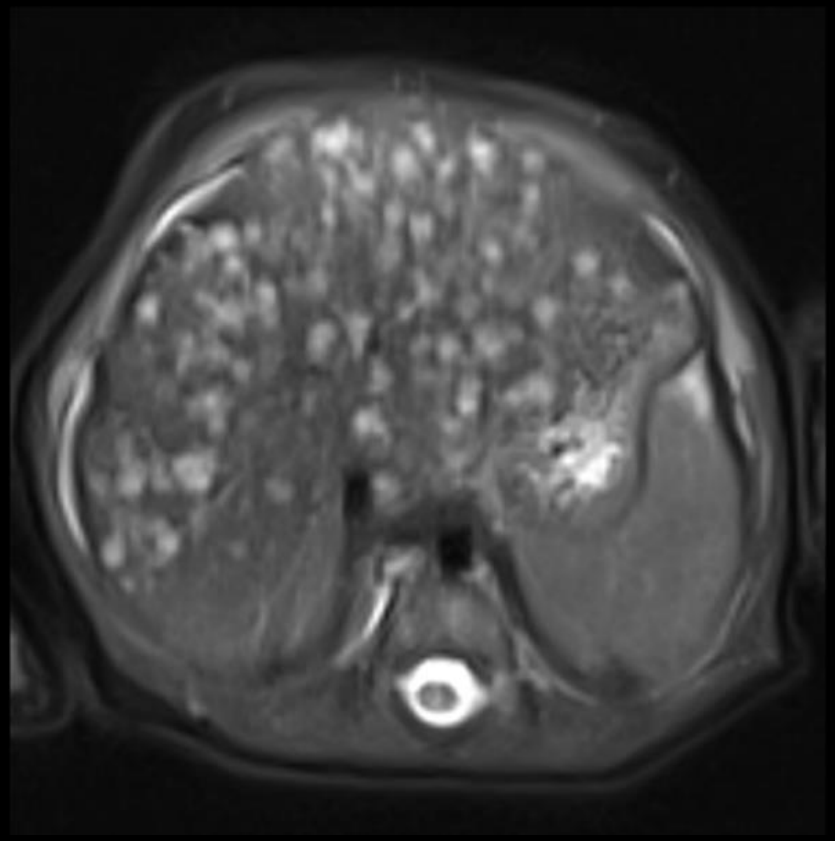
- CBC
  - WBC: 7.9, Hgb: 7.3 (L), Hct: 21.4 (L), PLT: 416
- CMP
  - Na: 139, K: 4.7, Cl: 109, CO2: 21, BUN: 10, Cr: 0.21, GLC: 110, Ca: 10.2
  - Total protein: 5.4, T. Bili: 0.4, AST: 42, ALT: 26, Alk Phos: 289, Albumin: 4.0
- Thyroid function tests
  - TSH: 5.393 (H), T4: 1.32
- Tumor markers
  - AFP: 785.2 (H)
  - Urinary vanillylmandelic acid (VMA) and homovanillic acid (HVA): Negative

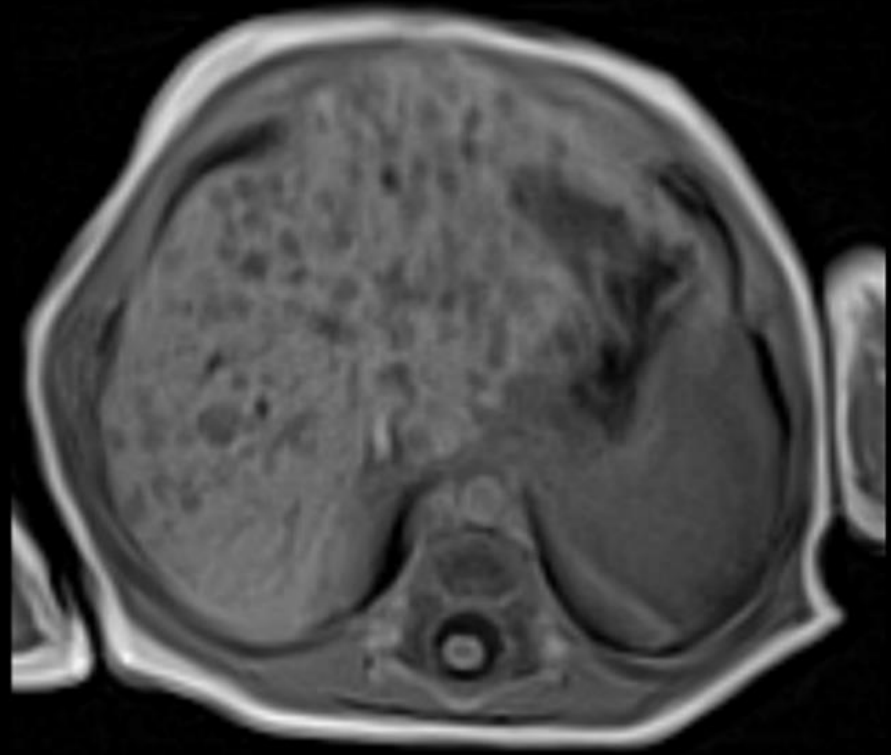
Physical Examination



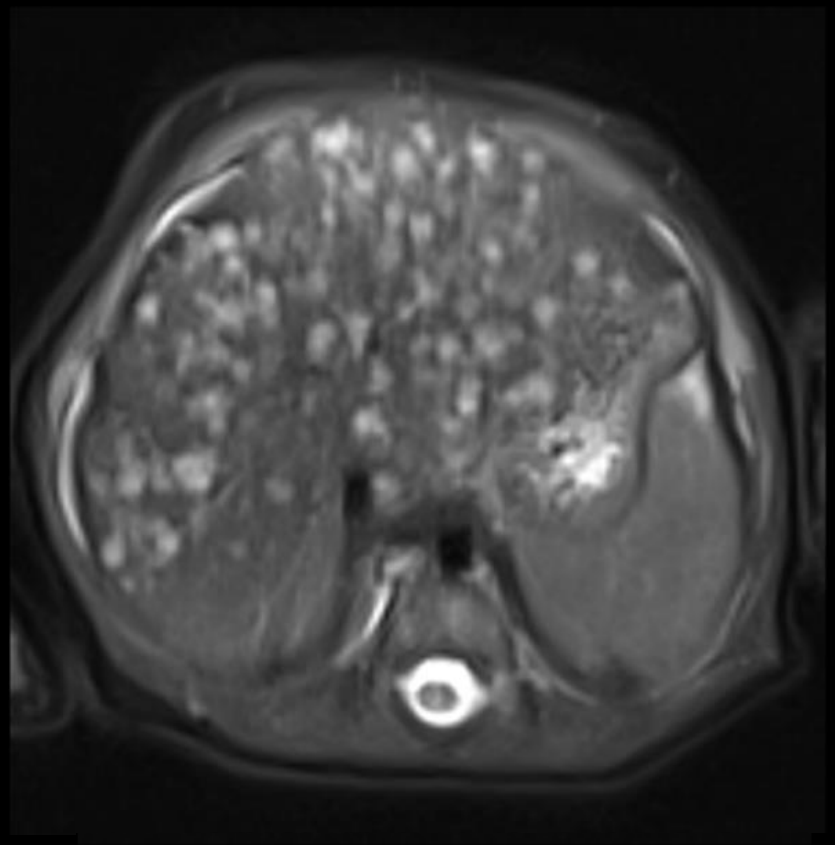


MRI Findings (unlabeled)

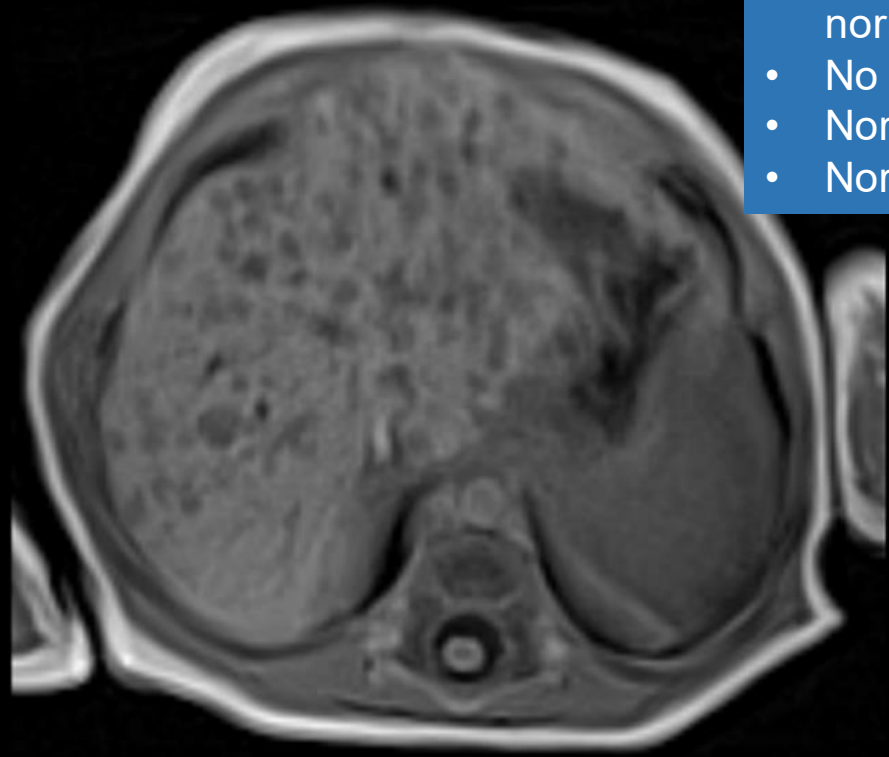




MRI Findings (labeled)



MRI of the liver: Extensive T2-hyperintense lesions



Lesions are hypointense on T1-weighted images

Pertinent negatives on MRI scan

- Adrenal glands were normal
- No paraspinal mass
- Normal bones
- Normal marrow signal

Final Dx:

Diffuse Infantile Hepatic Hemangioma

Case Discussion

- DIHH is rare (approximately 1/100,000)
- Pathogenesis is poorly understood
  - Possible dysfunctional response of pluripotent stem cells to hypoxia and RAAS
- Presence of GLUT1 (glucose transporter isoform 1) can help detect IHH and distinguish it from other vascular anomalies
- Most cases of DIHH are asymptomatic and are found incidentally on imaging
- Hepatomegaly is thought to be secondary to IVC compression by rapidly growing vascular masses

Case Discussion

- Anemia, thrombocytopenia, and consumptive coagulopathy can also result due to sequestering of RBCs, platelets, and coagulation factors within the tumors
- Hypothyroidism can result secondary to overproduction of iodothyronine deiodinase type 3 and thyrotropin-like factor
- While IHH is the most common diagnosis in infants <6 months with multiple hypoechoic liver lesions, metastatic disease (neuroblastoma most commonly) should also be considered, particularly in infants older than 6 months.

Case Discussion

- Treatment
  - Propranolol
    - 0.6mg/kg Propranolol BID initially, escalating to 1.7mg/kg Propranolol BID over two weeks with treatment sustained for 6-12 months based on clinical condition
      - Notably, during the tapering phase of treatment, rebound growth can be observed
    - Though the mechanism is poorly understood, propranolol has an antiproliferative effect in IHH2. Possible mechanisms for this include Vasoconstriction via pericytes, reduction in VEG-F and FGF and inactivation of the RAAS
    - Corticosteroids, Vincristine, and Levothyroxine have been used to supplement propranolol in the treatment of DIHH
  - Surgical or Interventional Radiology treatment
    - Indicated with life-threatening symptoms
    - Hepatic artery embolization or ligation - helps to counteract cardiac failure due to shunting
    - Orthoptic liver transplant - in DIHH since total hepatectomy is not a viable option
- Case outcome
  - The patient responded well to propranolol therapy, with rapid resolution of both the liver and the cutaneous hemangiomas.
  - Clinical follow-up at 1-year post-treatment showed a thriving, well developing child.

References:

1. Kulungowski AM, Alomari AI, Chawla A, Christison-Lagay ER, Fishman SJ. Lessons from a liver hemangioma registry: subtype classification. J Pediatr Surg (2012) 47(1):165–70. doi: 10.1016/j.jpedsurg.2011.10.037
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3. Shah SD, Baselga E, McCuaig C, Pope E, Coulie J, Boon LM, et al. Rebound growth of infantile hemangiomas after propranolol therapy. Pediatrics (2016) 137(4):e20151754. doi: 10.1542/peds.2015-1754
4. Yeh I, Bruckner AL, Sanchez R, Jeng MR, Newell BD, Frieden IJ. Diffuse infantile hepatic hemangiomas: a report of four cases successfully managed with medical therapy. Pediatr Dermatol (2011) 28(3):267–75. doi: 10.1111/j.1525-1470.2011.01421.x

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